FACULDADE DE MEDICINA
UNIVERSIDADE D
COIMBRA

MESTRADO INTEGRADO EM MEDICINA – TRABALHO FINAL

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Decompressive surgery in Chiari's malformation type 1: a retrospective analysis

ARTIGO CIENTÍFICO ORIGINAL

ÁREA CIENTÍFICA DE NEUROCIRURGIA

Trabalho realizado sob a orientação de:

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FEVEREIRO/2022

Decompressive surgery in Chiari's malformation type 1: a retrospective analysis

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Abstract

Introduction: The Chiari malformation type I (CM1) is a condition in which brain tissue extends below the foramen magnum and, sometimes, a syringomyelic cavity and hydrocephalus are present. This study aimed to associate decompressive surgery, by occipitopalatine line increase rate, with the “Chicago Chiari Outcome Scale” (CCOS) of the patient and the possibility to establish, in preoperative routines, a sagittal extension of bone to be removed, to obtain a desirable result, without complications, like pseudomeningocele (leakage of cerebrospinal fluid through a dural defect).

Material and Methods: We realized an observational and retrospective study with patients diagnosed with CM1, surgically treated at Coimbra Hospital and Universitary Centre. The syringomyelic cavity and occipitopalatine line were measured in computerized tomography scans or magnetic resonance imaging. Hydrocephalus was not found. The information for the CCOS was taken on the clinical files of the patients. For the statistical analysis, we performed a descriptive statistic, Kolmogorov-Smirnov test, Chi-square test of independence, the Eta Coefficient test and a receiver operating characteristic curve.

Results: After the surgery, the syringomyelic cavity decreased in all the patients. Cervical pain, cerebellar signs, alterations of sensitivity and motor signs were dependents on the presence of syringomyelia. Pseudomeningocele was more frequent in patients with syringomyelia (p=0.04). Besides that, occipitopalatine line increase rate was correlated with the presence of pseudomeningocele (p=0.001).

Discussion: The presence of syringomyelia influenced the clinical finds. There is no significant correlation between occipitopalatine line increase rate and CCOS. The development of pseudomeningocele is influenced by the presence of syringomyelia and is associated to a small increase of occipitopalatine line. So, removing a small amount of skull, it is more probable to have pseudomeningocele and, consequently, a worst outcome.

Conclusion: The amount of bone removed affects the surgical outcome. Not directly as expected, but indirectly because a small increase of occipitopalatine line is associated with the appearance of pseudomeningocele. Besides that, pseudomeningocele is more frequent in the group of patients who has syringomyelia. It is important to do better planning of surgery to avoid complications after it and to use the correct scores to follow-up the patient after surgery.

Keywords: “Syringomyelia”, “Chiari malformation”, “Occipitopalatine line”, “Chicago Chiari Outcome Scale”, “Decompressive surgery”.

Resumo

Introdução: A malformação de Chiari tipo 1 (CM1) é uma condição em que o tecido nervoso se estende além o foramen magnum, estando por vezes presentes uma cavidade siringomiélica e hidrocefalia. Este estudo tem como objetivo associar a cirurgia descompressiva com "*Chicago Chiari Outcome Scale*" (CCOS), bem como a possibilidade de se estabelecer, em rotinas pré-operatórias, uma extensão sagital de osso a remover, de modo a obter-se um resultado desejável, sem complicações, como pseudomeningocelo (extravasamento de líquido cerebroespinhal por um defeito na dura mater).

Material e Métodos: Realizamos um estudo observacional e retrospetivo com doentes diagnosticados com CM1, tratados cirurgicamente no Centro Hospitalar e Universitário de Coimbra. A cavidade siringomiélica e a linha occipitopalatina foram avaliadas através de tomografias computorizadas e ressonâncias magnéticas. Não foi encontrada hidrocefalia nos doentes. A informação para CCOS foi recolhida nos processos clínicos dos doentes. Para a análise estatística, realizamos uma estatística descritiva, teste Kolmogorov-Smirnov, teste Qui-quadrado de independência, teste coeficiente de Eta e curva de Característica de Operação do Receptor.

Resultados: Após a cirurgia, a cavidade siringomiélica diminuiu em todos os doentes avaliados. Cervicalgias, sinais cerebelosos, alterações na sensibilidade e sinais motores estavam na dependência da siringomielia. Pseudomeningocelo foi mais frequente nos doentes com siringomielia (p=0,04). Para além disso, a taxa de aumento da linha occipitopalatina estava relacionada com a presença de pseudomeningocelo (p=0,001).

Discussão: A presença de siringomielia influenciou os achados clínicos. Não há uma correlação significativa entre a taxa de aumento da linha occipitopalatina e CCOS. A presença de pseudomeningocelo é influenciada pela presença de siringomielia e está associada a um pequeno aumento da linha occipitopalatina. Por isso, ao remover-se pouco osso, é mais provável de se ter pseudomeningocelo e, consequentemente, um pior *outcome* cirúrgico.

Conclusão: A quantidade de osso removida afeta o *outcome* cirúrgico. Não diretamente como o esperado, mas indiretamente porque o pouco aumento da linha occipitopalatina está relacionado com o desenvolvimento de pseudomeningocelo. Para além disso, o pseudomeningocelo é mais provável em doentes com siringomielia. Revela-se importante executar um melhor planeamento da cirurgia de modo a evitar complicações e usar os melhores scores para realizar o follow-up dos doentes.

Palavras-chave: “Siringomielia”, “Malformação de Chiari”, “Linha occipitopalatina”, “Chicago Chiari Outcome Scale”, “Craniectomia descompressiva”.

Abbreviation

CCOS - “Chicago Chiari Outcome Scale”

CDT - Complementary Diagnostic Test

CHUC - Coimbra Hospital and Universitary Centre

CM1 - Chiari Malformation Type I

CSF – Cerebrospinal Fluid

CT Scans - Computerized Tomography Scans

FM - Foramen Magnum

MRI - Magnetic Resonance Imaging

PCF - Posterior Cranial Fossa

ROC curve - Receiver Operating Characteristic curve

SD – Standard Deviation

SPSS - “Statistical Package for the Social Sciences”

Introduction

The Chiari malformation type I (CM1) is a condition, congenital or acquired, on the posterior cranial fossa (PCF). In CM1, the PCF is small to room the hindbrain, affecting the circulation of the cerebrospinal fluid (CSF) and, sometimes, a syringomyelic cavity and hydrocephalus are present. Radiologically, CM1 may present a caudal herniation of one or both cerebellar tonsils through the foramen magnum (FM). The herniation is defined when the cerebellar tonsils are, at least, 3-5mm below the FM.[1]

The global incidence of CM1 has been cited as 0.24%-3.6% and 0.56%-0.77% in imaging exams.[2] CM1 is more prevalent in females.[2] About 30-70% of cases present syringomyelia[1-6] - a fluid cavity caused by disturbance of the normal CSF circulation. This enlargement of the central canal of spinal cord is more frequent in the cervical cord. Hydrocephalus is present in 7-10% of the patientes with CM1.[3, 7]

There are different symptoms and signs. The most common symptom is headache,[4, 8, 9] exacerbated by the Valsalva maneuver and neck extension. Cervical pain may exist ("thunder-like" pain), with arms irradiation. Vertigo and dizziness are also common. In the physical exam: nystagmus, "cape" sensory loss (dependent on the syringomyelia), motor signs, ataxia, muscular atrophy (also dependent on the syringomyelia),[8] cerebellar signs (dysmetria, ataxia, intention tremor, slurred speech, hypotonia), and dysesthesias may be present.[2, 3]

The majority of patients are treated surgically, when indicated.[10] The decompressive craniectomy of the PCF, with enlargement of FM, is the most common surgical treatment (**fig.1**). It is provided a more effective decompression realizing an augmentative duraplasty - with autologous or non-autologous tissue.[11-14]

The surgical outcomes are diverse and depend on several variables. The outcomes are affected by patient's age, patient's clinic, duration of symptoms, compression of nervous structures, the existence of syringomyelia, and decompressive craniectomy technique. Furthermore, the appearance of complications after surgery affects the patient's recovery. Postoperative pseudomeningocele and CSF fistulas stand out as two of many possible complications.[1, 15-19]

Postoperative pseudomeningocele is a surgical complication resulting from leakage of CSF through a dural defect, due to a rupture of arachnoid and dura mater. It can form a valve-way for the outflow of CSF to the surrounding tissues, wich can be enclosed by a fibrous capsule.[20-23]

Fig. 1 – Sagittal T2 pre-operative (1A) and post-operative (1B) MRI of a patient diagnosed with CM1 with syringomyelia. We can see a syringomyelic cavity bigger in fig.1A than in fig.1B.

Many scores were developed to better scale the outcomes of decompressive surgery. The “Chicago Chiari Outcome Scale” (CCOS) is one of the most complete scores.[24-27] The CCOS (**Table 1**) evaluates four parameters: pain symptoms, non-pain symptoms, functionality, and complications.[24] These parameters are evaluated from 1 to 4, for a minimum final score of 4 and a maximum final score of 16.

However, much remains to be defined in CM1. Despite having well-defined predictors of prognosis, it is not possible to correlate those predictors with the surgery, in order to anticipate the surgical result. There is a lack of craniometric references for decompressive surgery. Besides, previous studies have failed to establish a coherent relationship between increased PCF and the effectiveness of surgery. It is not known the ideal amount of bone to remove in order to achieve a favourable surgical outcome.

Consequently, it is necessary to know how decompressive craniectomy influences the evolution of the pathology. This study proposes to associate the decompression of nervous structures with the improvement of symptoms, according “Chicago Chiari Outcome Scale”. For this purpose, occipitopalatine line (defined as a line that connects the posterior point of the hard palate to the most caudal point of the occipital curve, before and after surgery) helps us to objectify the clinical and imaging improvements.[28-31] Finally, we evaluate the complications of the surgery, specially the pseudomeningocele. In a retrospective observational study, we inferred the relevance of this surgical procedure, evaluating its effectiveness in the context of this pathology. In

this way, it will be possible to establish, in preoperative routines, an ideal amount of bone to be removed in order to obtain a desirable and expected result, without complications.

Table 1 – "*Chicago Chiari Outcome Scale*"[24]

Pain	*Non-pain*	*Functionality*	*Complications*
1-Worse	1- Worse	1-Unable to attend	1-Persistent complication, poorly controlled
2-Unchanged and refractory to medication	2- Unchanged and refractory to medication	2-Moderate impairment (<50% attendance)	2-Persistent complication, well controlled
3-Improved or controlled with medication	3- Improved or controlled with medication	3-Mild impairment (>50% attendance)	3-Transient complication
4-Resolved	4-Resolved	4-Fully functional	4-Uncomplicated course

Adapted from "Aliaga L, Hekman KE, Yassari R, Straus D, Luther G, Chen J, et al. A novel scoring system for assessing Chiari malformation type I treatment outcomes. Neurosurgery. 2012;70(3):656-64; discussion 64-5. doi: 10.1227/NEU.0b013e31823200a6. PubMed PMID: 21849925; PubMed Central PMCID: PMC6718293."

Material and Methods

Study design

We realized an observational, retrospective and analytical study. The population studied were patients diagnosed with CM1, surgically treated at Coimbra Hospital and Universitary Centre (CHUC) between 2005 and 2019. We admitted patients diagnosed with CM1, who underwent decompressive surgery of PCF and who did not require ventricular shunt.

The clinical files and the complementary diagnostic tests (CDT) of the patients were analyzed. The approval of the CHUC's ethics committee was obtained.

The bibliographic search was carried out through the PubMed database, associating multiple keywords: “Chiari malformation”, “Decompressive surgery”, “Chicago Chiari Outcome Scale”, “Syringomyelia”, “Hydrocephalus”, “Pseudomeningocele”, “McGregor’s line”, “McRae’s line”. Additional research was carried out through the Greenberg M, “*Handbook of Neurosurgery*”, 9th edition, 2020.

Procedure

- From May to June: structuring and planning the study;
- July: request the approval of the CHUC's ethics committee (appendix I);
- From October to November: analyzing the clinical files and CDT of the patients and elaboration of the database of the study;
- From November to February: Statistical analysis of the data and writing the study.

Database explanation

The patients were separated into two groups: patients diagnosed with CM1 with syringomyelia, and patients diagnosed with CM1 without syringomyelia.

In the group of patients diagnosed with CM1 without syringomyelia, we considered the following parameters: gender, age, patient's clinic, physical exam, surgery's date, age on surgery's date, evolution after surgery, complications, grade of cerebellar tonsillar herniation, length of occipitopalatine line before surgery, length of occipitopalatine line after surgery, occipitopalatine line increase rate.

In the group of patients diagnosed with CM1 with syringomyelia, we also considered the same parameters. Additionally, we considered: in a sagittal plane, the maximum diameter of the syringomyelic cavity before and after surgery.

Hydrocephalus was not found in both groups.

The age of the patients was divided into 3 groups: less than 40 years old, between 40 and 60 years old and more than 60 years old. These were defined as “age groups”.

Time of follow up, minimum and maximum, was 3 months and 24 months, respectively.

About CDT, we prioritized pre- and postoperative magnetic resonance imaging (MRI).(32) In the group of patients diagnosed with CM1 without syringomyelia, we realized the following measurements:

- Grade of cerebellar tonsillar herniation, in millimetres, below McRae's line[30, 31] - a line drawn in a midsagittal section that connects the anterior and the posterior margins of the FM (*Basion* to *Opisthion*) (Fig.2);
- Length, in millimetres, of occipitopalatine line before surgery - a line drawn in a midsagittal section that connects the posterior point of the hard palate to the most caudal point of the occipital curve (similar to McGregor’s line)[30, 31] (Fig.3A);
- Length, in millimetres, of occipitopalatine line after surgery - a line drawn in a midsagittal section that connects the posterior point of the hard palate to the most caudal point of the remnant occipital curve (an extension of McGregor’s line)[30, 31] (Fig.3B);

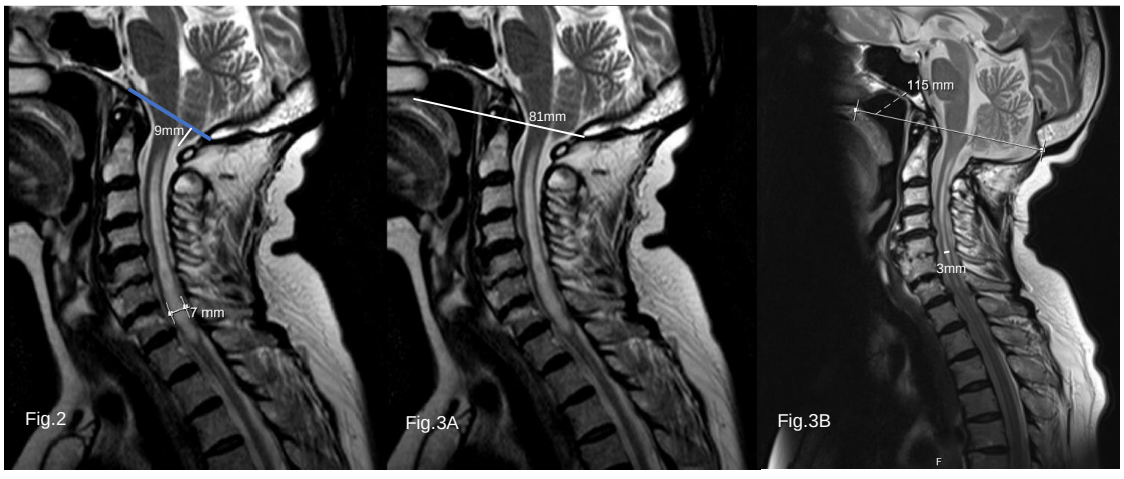

Fig.2 - Mid-sagittal T1 MRI of a patient diagnosed with CM1 with syringomyelia. The blue line indicates McRae's line. The grade of cerebellar tonsillar herniation, in millimeters, is measure below McRae's line (9mm). Sagittal diameter of the syringomyelic cavity before surgery was 7mm. **Fig.3A** - Mid-sagittal T1 MRI of the same patient. The white line indicates the length, in millimeters, of occipitopalatine line before surgery (81mm). **Fig.3B** - Mid-sagittal T2 MRI of the same patient. The white line indicates the length, in millimeters, of occipitopalatine line after surgery (115mm). Sagittal diameter of the syringomyelic cavity after surgery was 3mm.

To calculate occipitopalatine line increase rate, we used the following equation:

$$\frac{\left(\begin{array}{c}\text{length of occipitopalatine}\\ \text{line after surgery}\end{array}\right) - \left(\begin{array}{c}\text{length of occipitopalatine}\\ \text{line before surgery}\end{array}\right)}{\text{length of occipitopalatine line before surgery}} \; x\,100$$

After that, we divided all the results into three groups: 0%-24% as a "Small increase"; 24%-43% as a "Moderate increase"; and >43% as a "Large increase". To achieve that, we tested multiple cut-off points, inclusive cut-off points at the standard deviation (SD) of the normal distribution. The best results were obtained with a cut-off at 24% and with a cut-off at 43%. The last one was obtained by mean + 1SD (32% + 11%, respectively).

In the group of patients diagnosed with CM1 with syringomyelia, we also considered the same measurements. Additionally, we realized the following measurements:

- Sagittal diameter of the syringomyelic cavity before surgery;
- Sagittal diameter of the syringomyelic cavity after surgery.

In the absence of MRI, we analyzed pre- and postoperative computerized tomography scans (CT scans). Inclusively, the report of the imaging exams, where we could find, in more detail, the grade of cerebellar tonsillar herniation, in millimeters, and the progression of the syringomyelic cavity.

The grade of herniation was divided into two groups: those with 3-5mm below the FM and the others with more than 5mm.

For both groups (with and without syringomyelia), we calculated the surgical outcomes by using the CCOS. We looked at the progression after surgery and complications. To calculate the CCOS, we used the information in the clinical files collected after the decompressive craniectomy. Patients were evaluated according to pain symptoms, non-pain symptoms, functionality (capacity to attend work and to realize daily activities), and postoperative complications, getting a score from 1 to 4 in each parameter. All results were integrated in postoperative CCOS.

Pain symptoms usually included headaches, shoulder, neck, arms, and legs pain. If these symptoms are completely resolved after the surgery, the patient received a 4. New postoperative pain symptoms were not considered (for example, neck pain trauma related to Chiari surgery). If the pain improved, with or without medications, the patient received a 3. That means that the surgery decreases the pain in intensity, duration, and/or frequency, even without full resolution. When the pain symptoms were unchanged

and refractory to medication, the patients received a 2. However, if the pain worsened, the patient received a score of 1.

Non-pain symptoms usually included sensory changes/loss, muscular atrophy, vertigo, dizziness, dysphagia, ataxia, and other neurological signs. The grading strategy is very similar to the grading strategy of the pain symptoms. If these symptoms are completely resolved after the surgery, the patient received a 4. If these symptoms improved, with or without medications, the patient received a 3. When the non-pain symptoms were unchanged and refractory to medication, the patients received a 2. However, if the non-pain symptoms worsened, the patient received a score of 1.

Patients without symptoms (pain and/or non-pain symptoms) got a score of 4 in each one of the affected groups.

Functionality is defined as the capacity to attend work and to realize daily activities. When the patient was able to attend to all the duties received a score of 4. If there was any impairment, greater than 50%, the patient received a 3. Less than 50%, the patient received a 2. When the patient was unable to attend their duties at all received a 1.

Postoperative complications included CSF leaks, wound infections, meningitis, and CSF obstruction. If there are no complications, the patient received a score of 4. When the patient experienced a transient complication, like a wound infection, meningitis, or a transient increased of the intracranial pressure, the patient received a 3. When occurred a persistent complication that was well-controlled, the patient received a score of 2. For example, pseudomeningocele that is stable and well-controlled led to a score of 2. However, if the pseudomeningocele cannot be well-controlled, the patient received a 1. Also, the patient received a score of 1 if experienced any other persistent complications that were poorly controlled, medically and/or surgically.

The final score was between 4 and 16 and were divided into two groups: "Good outcome" and "Bad outcome". We established the cut-off between 4-12 ("Bad outcome") and 13-16 ("Good outcome").[24] Sensitivities and specificities were calculated to determine the best cut-off point for this division. A receiver operating characteristic (ROC) curve (Fig.4) was performed between the CCOS with headache improvement and the area under the ROC curve was 0.703. These results showed that a CCOS of 13 has a good sensitivity and specificity to identify a “Good outcome".

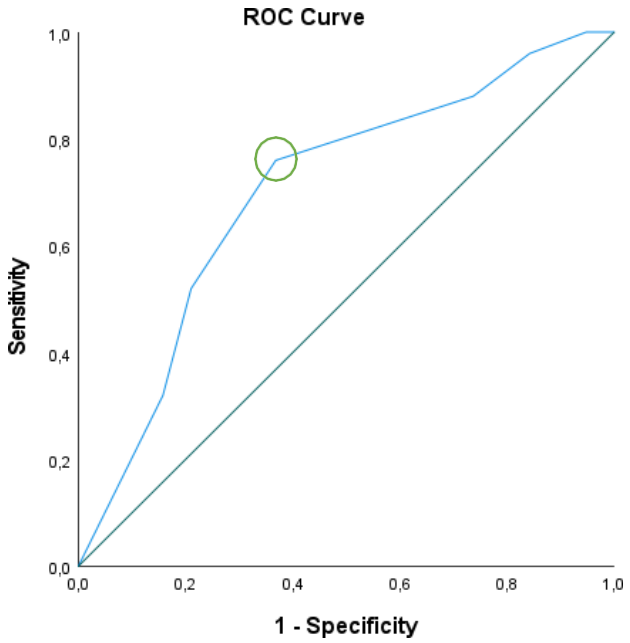

Fig.4 - Graphic showing the ROC curve for prediction of CCOS using headache improvement. The area under the curve is 0.703 (p<0.001).

Data analysis

For the statistical analysis was used the "Statistical Package for the Social Sciences" (SPSS) software, version 27, with a significance level of 0.05.

Descriptive statistics were performed considering mean, standard deviation and valid percent. The normality test used was *Kolmogorov-Smirnov test* and the comparison between groups was performed with *Chi-square test of independence*. The *Eta Coefficient test statistic* was used to verify an association between gender and age on surgery's date (minimum level for acceptance: 0.2).

To determine both groups of CCOS, a ROC curve was used to predict the cut-off of CCOS using headache improvement.

Results

We analyzed 61 patients diagnosed with CM1 and treated with a decompressive craniectomy of the PCF, between 2005 and 2019, at CHUC. The age on surgery's date of the patients was between 18 and 72 years old. The mean age on surgery's date was 48.59 +/- 12.16 years old and the median age was 52 years old. 17 patients (27.9%) were under 40 years old, 36 patients (59%) between 40 and 60 years old, and 8 patients (13.1%) above 60 years old.

We had 14 male patients (23% of the total) - mean age was 41.79 +/- 12.95 years old and the median age was 39 years old. And 47 female patients (77%) - mean age was 50.62 +/- 11.28 years old and the median age was 53 years old. The *Eta Coefficient test statistic* (η) is 0.307, which determined an association between gender and age on surgery's date.

Syringomyelia was present in 23 patients (37.7%). 8 patients were men (34.8%) and 15 patients were women (65.2%). The mean age was 48.13 +/- 14.45 years old and the median age was 53 years old. Minimum of 18 and maximum of 72.

In the patients without syringomyelia, 6 patients were men (15.8%) and 32 patients were women (84.2%). The mean age was 48.87 +/- 10.751 years old and the median age was 51 years old. Minimum of 25 and maximum of 72.

Due to lack of imaging exams, the grade of cerebellar tonsillar herniation was determinated only in 45 patients (73.77%), in which 40 patients had an herniation more than 5mm below the FM.

About the clinical findings, the results are shown in fig.5:

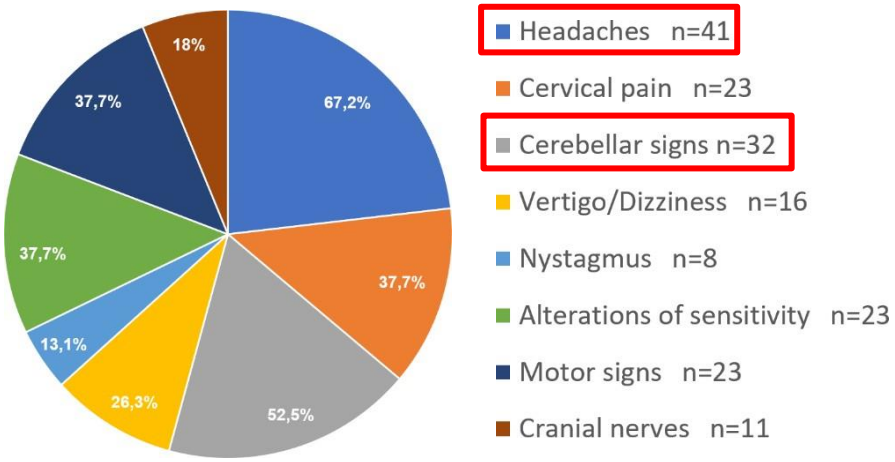

Fig. 5 – Frequency of symptoms and signs in all the patients. The most frequent symptoms and signs are in a red rectangle. About cerebellar signs, we considered dysmetria, ataxia, intention tremor, slurred speech and hypotonia.

Around 39% of the patients with headaches have an exacerbation with Valsalva maneuver. The most commom cranial nerves affected was IX and X (decrease/absence of gag reflex was the most frequent finding).

Due to lack of enough information, it was only possible to calculate the CCOS ("Chicago Chiari Outcome Scale") of 44 patients. The results were as follows: minimum of 4 and maximum of 16; the mean was 13.68 +/- 2.31 and the median was 14. 8 patients (18.2%) had a "Bad outcome" (range 4-12) and 36 patients (81.8%) had a "Good outcome" (range 13-16). This variable does not have a normal distribution, according to the *Kolmogorov-Smirnov one-sample test* (p<0.001).

One fatality was reported among the 61 patients. A female patient was diagnosed with CM1 with syringomyelia and with a grade of cerebellar tonsillar herniation >5mm. After surgery, she developed many complications: extra-axial hematoma, a CSF leakage and hydrocephalus that needed a ventriculoperitoneal shunt. She underwent 2 more decompressive surgeries: 2 and 3 months after the first intervention. Her clinical condition deteriorated, needed advanced support of life and died. Because of this, her CCOS was 4.

Only in 39 patients was possible to calculate occipitopalatine line increase rate, completing our database. So, our eligible number of patients was 39 (63.9% of the patients). We can affirm, with 95% of confidence, that the mean of the population is between 28.4797% and 35.6118%. 8 patients (20.5%) had a "Small increase", 23 patients (59.0%) had a "Moderate increase" and 8 patients (20.5%) had a "Large increase". This variable has a normal distribution (**Table 2**), according to the *Kolmogorov-Smirnov one-sample test* (p>0.05).

Table 2 – Normality test of occipitopalatine line increase rate

	Kolmogorov-Smirnov[a]			**Shapiro-Wilk**		
Occipitopalatine line increase rate	Statistic	df	Sig.	Statistic	df	Sig.
	0.097	39	**0.200**	0.975	39	0.511

In line with the results, *Chi-square nonparametric statistic* was used.

Comparison between the gender and clinical finds:

A correlation was found between the female patients and headaches. In others clinical finds, there is a tendency in vertigo/dizziness and nystagmus (**Table 3a**). We looked for the *Fisher's Exact Test* because 1 cell (25.0%) have expected count less than 5.

Table 3a – *Chi-square test* between gender and clinical finds

	Clinical find		**Headaches**	**Vertigo/ Dizziness**	**Nystagmus**
Male (*n*=14)	Yes	Count	6	1	0
		Expected count	9.4	3.7	1.8
	No	Count	8	13	14
		Expected count	4.6	10.3	12.2
Female (*n*=47)	Yes	Count	35	15	8
		Expected count	31.6	12.3	6.2
	No	Count	12	32	39
		Expected count	15.4	34.7	40.8
		Chi-square	4.891	3.421	2.743
		dF	1	1	1
		Asymp. Sig.	0.027	0.064	0.098
		Fisher's Exact Test	**0.049**	0.088	0.18

Comparison between the presence of syringomyelia and clinical finds:

The presence of syringomyelia is correlated with: headaches (*p*=0.012), cervical pain (*p*=0.018), alterations of sensitivity (*p*=0.001) and motor signs (*p*=0.001) (**Table 3b**). We looked for *Asymp. Sig.* because 0 cells (00.0%) have expected count less than 5.

Table 3b - *Chi-square test* between the presence of syringomyelia and clinical finds

	Clinical find		**Headache**	**Cervical Pain**	**Cerebellar signs**	**Alteration sensitivity**	**Motor Sings**
With syring.[a] (*n*=23)	Yes	Count	11	13	15	15	15
		Expected count	15.5	8.7	12.1	8.7	8.7
	No	Count	12	10	8	8	8
		Expected count	7.5	14.3	10.9	14.3	14.3
Without syring.[b] (*n*=38)	Yes	Count	30	10	17	8	8
		Expected count	25.5	14.3	19.9	14.3	14.3
	No	Count	8	28	21	30	30
		Expected count	12.5	23.7	18.1	23.7	23.7
		Chi-square	6.297	5.566	2.41	11.898	11.898
		dF	1	1	1	1	1
		Asymp. Sig.	**0.012**	**0.018**	0.121	**0.001**	**0.001**
		Fisher's Exact Test	0.023	0.029	0.186	0.001	0.001

a. Patients diagnosed with CM1 with syringomyelia

b. Patients diagnosed with CM1 without syringomyelia

Comparison between grade of cerebellar tonsillar herniation and clinical finds:

A tendency between grade of cerebellar tonsillar herniation and two clinical finds was discovered: cerebellar signs and the alteration of sensitivity (**Table 3c**). We looked for the *Fisher's Exact Test* because 5 cell (50.0%) have expected count less than 5.

Table 3c - *Chi-square test* between grade of cerebellar tonsillar herniation with the clinical finds

	Clinical find		**Cerebellar signs**	**Alteration of sensitivity**
≤5[a] (*n*=5)	Yes	Count	1	4
		Expected count	2.6	2
	No	Count	4	1
		Expected count	2.4	3
>5[b] (*n*=40)	Yes	Count	22	14
		Expected count	20.4	16
	No	Count	18	26
		Expected count	19.6	24
		Chi-square	2.179	3.75
		dF	1	1
		Asymp. Sig.	0.14	0.053
		Fisher's Exact Test	0.187	0.141

a. Grade of herniation of 5mm or less below the FM.

b. Grade of herniation more than 5mm below the FM.

Comparison between the presence of syringomyelia with gender and grade of cerebellar tonsillar herniation:

There is no correlation between the presence of syringomyelia and both topics. Nevertheless, a tendency between these two Chi-square tests was found (**Table 3d**). We looked for *Asymp. Sig.* because 0 cells (00.0%) have expected count less than 5.

Table 3d - *Chi-square test* between the presence of syringomyelia with gender and grade of cerebellar tonsillar herniation

			Gender (*n*=61)		Tonsils herniation (*n*=45)	
			Male	**Female**	**≤5**	**>5**
With syring?[a]	Yes	Count	8	15	4	14
		Expected count	5.3	17.7	2	16
	No	Count	6	32	1	26
		Expected count	8.7	29.3	3	24
		Chi-square	2.875		3.75	
		dF	1		1	
		Asymp. Sig.	0.087		0.052	
		Fisher's Exact Test	0.119		0.141	

a. Selected cases of patients diagnosed with CM1 with syringomyelia

Comparison between the CCOS with occipitopalatine line increase rate, age groups and if the pain symptoms improved:

We did not find any correlation between occipitopalatine line increase rate and age groups with the CCOS. In these two *Chi-square tests of independence*, we looked for the *Monte Carlo Sig.* because we had more than two variables (**Table 3e**).

Table 3e - *Chi-square test* between the CCOS with occipitopalatine line increase rate, age groups and if the pain symptoms improved

		Occipitopalatine line increase rate (*n*=39)		
		Small increase	**Moderate increase**	**Large increase**
Bad outcome[a]	Count	2	5	0
	Expected count	1.4	4.1	1.4
Good outcome[b]	Count	6	18	8
	Expected count	6.6	18.9	6.6
	Chi-square	2.244		
	dF	2		
	Asymp. Sig.	0.326		
	Monte Carlo Sig.	0.513		
	Fisher's Exact Test			

For this analysis, we had not the imaging exams of all the 44 cases.

a. Classified as “Bad outcome” in CCOS

b. Classified as “Good outcome” in CCOS

		Age Groups (*n*=44)		
		<40	**40-60**	**>60**
Bad outcome[a]	Count	2	6	0
	Expected count	2.2	4.9	0.9
Good outcome[b]	Count	10	21	5
	Expected count	9.8	22.1	4.1
	Chi-square	1.426		
	dF	2		
	Asymp. Sig.	0.49		
	Monte Carlo Sig.	0.64		
	Fisher's Exact Test			

a. Classified as “Bad outcome” in CCOS

b. Classified as “Good outcome” in CCOS

		Pain symptoms improved? (*n*=36)	
		Yes	**No**
Bad outcome[a]	Count	3	4
	Expected count	4.9	2.1
Good outcome[b]	Count	22	7
	Expected count	20.1	8.9
	Chi-square	2.895	
	dF	1	
	Asymp. Sig.	0.089	
	Monte Carlo Sig.		
	Fisher's Exact Test	0.167	

a. Classified as “Bad outcome” in CCOS

b. Classified as “Good outcome” in CCOS

About the pain symptoms improvement, we did not find any correlation with CCOS, even if there was some tendency in that analysis. We looked for the *Fisher's Exact Test* because 2 cells (50.0%) have expected count less than 5 (**Table 3e**).

Comparison between gender with the presence of pseudomeningocele after the surgery:

We found a tendency for man to develop pseudomeningocele after surgery. We looked for the *Fisher's Exact Test* because 1 cell (25.0%) have expected count less than 5 (**Table 3f**).

Table 3f – *Chi-square test* between gender and pseudomeningocele after surgery

	Clinical find		**Pseudomeningocele**
Male (*n*=14)	Yes	Count	6
		Expected count	3.4
	No	Count	8
		Expected count	10.6
Female (*n*=47)	Yes	Count	9
		Expected count	11.6
	No	Count	38
		Expected count	35.4
		Chi-square	3.270
		dF	1
		Asymp. Sig.	0.071
		Fisher's Exact Test	0.087

Comparison between syringomyelia with the presence of pseudomeningocele after the surgery:

The presence of syringomyelia is correlated with the presence of pseudomeningocele after the surgery (*p*=0.04). We looked for *Asymp. Sig.* because 0 cells (00.0%) have expected count less than 5 (**Table 3g**).

Table 3g - *Chi-square test* between syringomyelia and pseudomeningocele after surgery

	Clinical find		**Pseudomeningocele**
With syring.[a] (*n*=23)	Yes	Count	9
		Expected count	5.7
	No	Count	14
		Expected count	17.3
Without syring.[b] (*n*=38)	Yes	Count	6
		Expected count	9.3
	No	Count	32
		Expected count	28.7
		Chi-square	4.209
		dF	1
		Asymp. Sig.	**0.04**
		Fisher's Exact Test	0.065

a. Patients diagnosed with CM1 with syringomyelia

b. Patients diagnosed with CM1 without syringomyelia

Comparison between occipitopalatine line increase rate with the presence of pseudomeningocele after the surgery:

We found an association (*p*=0.001) between occipitopalatine line increase rate with the presence of pseudomeningocele after the surgery. We looked for the *Monte Carlo Sig.* because we had more than two variables (**Table 3h**).

Table 3h - *Chi-square test* between occipitopalatine line increase rate with the presence of pseudomeningocele after the surgery

			Occipitopalatine line increase rate (*n*=39)		
			Small increase	**Moderate increase**	**Large increase**
Pseudo-meningocele	Yes	Count	7	4	1
		Expected count	2.5	7.1	2.5
	No	Count	1	19	7
		Expected count	5.5	15.9	5.5
		Chi-square	15.273		
		dF	2		
		Asymp. Sig.	<0.001		
		Monte Carlo Sig.	**0.001**		

a. Selected cases of patients with pseudomeningocele after surgery

We looked for the same comparison but only in the patients diagnosed with CM1 with syringomyelia. We selected cases of patients diagnosed with CM1 with syringomyelia and applied the same *Chi-square test* (**Table 3i**). We found an association (*p*=0.003) between occipitopalatine line increase rate with the presence of pseudomeningocele after the surgery in patients diagnosed with CM1 with syringomyelia. We looked for the *Monte Carlo Sig.* because we had more than two variables

Table 3i - *Chi-square test* between occipitopalatine line increase rate with the presence of pseudomeningocele after the surgery, only in patients diagnosed with CM1 with syringomyelia.

			Occipitopalatine line increase rate (*n*=19)		
			Small increase	**Moderate increase**	**Large increase**
Pseudo-meningocele	Yes	Count	6	1	1
		Expected count	2.5	3.8	1.7
	No	Count	0	8	3
		Expected count	3.5	5.2	2.3
		Chi-square	12.277		
		dF	2		
		Asymp. Sig.	0.002		
		Monte Carlo Sig.	**0.003**		

a. Selected cases of patients diagnosed with CM1 with syringomyelia, who developed pseudomeningocele after surgery

Syringomyelia's size evolution after surgery

In 15 patients with syringomyelia, after surgery, the cavity has downsized:

Table 4 – Frequency of symptoms and signs in all the patien

Downsizing of syringomyelia				
		Frequency	Percent	Valid Percent
Valid	Yes	**15**	65.2	**100.0**
Missing	System	8	34.8	
Total		23	100.0	

a. Measurement of the maximum sagittal diameter of the syringomyelic cavity, before and after the surgery.

Discussion

Our work focused on patients diagnosed with CM1 who needed surgery. Only the patients with symptoms or with syringomyelia undergone surgery.

In our sample, CM1 was more frequent in the 5th decade of life, and it was more frequent in female patients. The symptoms were more evident in female patients. The most frequent symptom was headache, which can be worsened by Valsalva maneuver.[9] Headache was more common in female patients - both variables correlated significantly (p=0.049). Although there was no significant relationship between the other symptoms, there was a tendency in some symptoms, like vertigo/dizziness. None male patients had nystagmus. Furthermore, age on surgery's date depended on gender - male patients were 9 years younger (on average) than female patients.

In other words, gender influenced the signs and symptoms of the disease. Symptoms (headache, dizziness, etc.) were more evident in women. It can explain the greater demand for medical help by female patients, with a consequent greater number of women diagnosed.[2] We can not deny the same prevalence in man just because we do not have the same clinical expression. As we will not perform imaging exams for all people, plus the less demand for help from men, the incidence in females seems to be higher.

The grade of cerebellar tonsillar herniation influenced the clinical presentation of the disease: cerebellar signs and alterations of sensitivity. The higher the grade of herniation (≥5mm below the FM), the more evident were the cerebellar signs - it can be explained by the caudal location of the cerebellum, in a smaller place than PCF, which compresses the vestibular fibers.

We did not find any correlation between the grade of cerebellar tonsillar herniation and syringomyelia.[33] Usually, to better define as CM1 in adults, it needs a grade of herniation ≥5mm below the FM.[1, 3] However, some patients have less than 5mm and need surgery too. And one reason for that was the existence of syringomyelia. On one hand, the symptoms of some patients are justified by the syringomyelic cavity, even with a smaller grade of herniation. On the other hand, the symptoms are justified by a grade of herniation ≥5mm below the FM, without any cavity.

The existence of syringomyelia did not change significantly the mean age compared to the group without syringomyelia. The mean age in the syringomyelia group

was 48.13, like the group without syringomyelia (48.87). In other words, the existence of syringomyelia did not motivate an earlier diagnosis.

In the group without syringomyelia, the most frequent gender was female (84.2%, vs 65.2%) and the most prevalent symptom was headache. The grade of cerebellar tonsillar herniation was higher in this group of patients than in the syringomyelia group. The grade of herniation was responsible for the symptoms, and it was the factor that led to the surgical decision.

In our study, syringomyelia is predominant in males. More than 50% of male patients (8 out of 14 male) had syringomyelia, against one third of female patients (15 out of 47 female). Some clinical finds were correlated with the existence of syringomyelia[25] - cervical pain, alterations of sensitivity, and motor signs. On the other hand, headaches were less frequent in the group of patients with syringomyelia.

The surgical approach of CM1 has a lack of information about the amount of bone to be removed. Decompressive surgery is performed by removing a random amount of skull to increase FM. We do not know if we are removing unnecessary bone or if we are leaving some bone to be removed. Some complications can happen in those two situations: the patient can have a caudal herniation[34, 35] of the brain if we remove too much skull; or, if we left some to remove, we do not solve the problem and a second surgery may be necessary.[36]

To have an adequate decompressive craniectomy is essential to consider the ideal amount of bone to be removed to enlarge FM. We evaluated, retrospectively, the decompressive craniectomy with the occipitopalatine line. After surgery, the result needs to be evaluated. For that, we used the “Chicago Chiari Outcome Scale” - CCOS[24-27] - a scale that allows differentiate those patients with a favourable or unfavourable outcome.

This evaluation can be performed at follow-up appointments, months after surgery, not immediately after, to increase its acuity. Even without being used in appointments, it provides important and systematized information on the patient's evolution, allowing a better interdisciplinary dialogue.

It would be logical to think that the best surgical outcome would be given by larger increase of occipitopalatine line because the surgery would give a wide space for PCF structures. However, the results showed that there is no correlation between occipitopalatine line increase rate and CCOS. Nevertheless, the results showed that all the patients with a large increase of occipitopalatine line had a good outcome. On the other hand, a small increase rate had a less favourable outcome.

One of the parameters of CCOS is the postoperative complications - to highlight the pseudomeningocele.[16-19] The results showed that a small decompressive craniectomy was correlated with the existence of pseudomeningocele (p=0.001). It means that it was not given enough space for an efficient decompression of PCF, limiting the circulation of CSF. This limitation explains the higher number of patients with pseudomeningocele in the syringomyelia group. Despite that, the syringomyelic cavity reduced in all patients, which demonstrates the effectiveness of the surgery in the treatment.

In this perspective, a smaller occipitopalatine line increase rate is related to a worse surgical outcome because it increases the probability to develop pseudomeningocele. In other words, if a larger increase rate were performed, it could prevent pseudomeningocele as complication. This increase rate arises as a craniectomy extension marker to enhance the surgical outcome.

Indirectly, the CCOS is dependent on occipitopalatine line increase rate. It is important to do a proper evaluation before surgery, to get the best result after it, by estimating that increase rate.

Pseudomeningocele was more frequent in male patients, and it was more frequent in the group of patients with syringomyelia. The existence of a syringomyelic cavity was a preditive factor of a worse outcome - a small increase rate got complicated with pseudomeningocele. All the patients in syringomyelia group who removed a little portion of occipital bone developed pseudomeningocele. Did not happen the same in the patients that removed a greater percentage of bone. Considering that a large increase in occipitopalatine line prevents the development of pseudomeningocele, the patients with syringomyelia need to remove a greater amount of bone, to enhance a good surgical outcome.

A close relationship was evident between males, CM1 with syringomyelia, and postoperative pseudomeningocele.

After the surgery, the syringomyelia has downsized in all the patients.[4, 5, 37, 38] Doing a decompressive surgery gives more space to the PCF structures, helping the CSF to flow properly. It is important to highlight this information to demonstrate that decompressive surgery is effective.

For a better evaluation of the PCF, a three-dimensional volumetric study could be used instead of the occipitopalatine line assessment. Even a bi-dimensional study, by evaluating the increase of the FM's coronal diameter, would be better, but it was not

possible because of the lack of imaging exams, particularly in axial planes. So, we tried a more simplistic, but solid, approach with less waste of human resources and time saver. This work may provide a foundation for future prospective studies in which a three-dimensional volumetric evaluation of the PCF may be performed, before and after surgery, and compared with the surgical outcome of the patient, by using the CCOS.

Being a retrospective study was the biggest limitation of the work. Besides, the absence of many of the imaging exams was another limitation: lack of CT scans and MRI, particularly in axial planes to determine the increase of the FM's coronal diameter. Despite that, some clinical information was poor and the appointments after surgery had not a complete check-up. That was the reason our eligible population was 39 out of 61 patients – because only these 39 patients had the information to make all the comparison studied. There was always subjectivity in analyzing the patients in the follow-up appointments. Plus, there is to mention the subjectivity of the investigators on analyzing those clinical files and all the imaging exams to formulate the CCOS and to calculate occipitopalatine line increase rate.

Conclusion

The CM1 can go unnoticed. Once it gives symptoms, it could be very harmful to the patient. With the radiological exams, the diagnosis is easier and favours a good preoperative routine. It is crucial to do that because the incidence is greater in the working-ages, and it is expected the best result after surgery.

Calculating occipitopalatine line increase rate could be a useful tool to help to reach the best result after surgery. Indirectly, removing a small amount of bone lead to a worst outcome. By removing a small amount of bone (a small increase of occipitopalatine line), the CSF does not have enough space to flow properly. It can complicate with pseudomeningocele, deteriorating the surgical outcome of the patient.

To prevent that, making some craniometric references by calculating occipitopalatine line increase rate seems to be beneficial. It is easy, does not take much time, and does not need extra exams. Thus, a moderate/large increase of occipitopalatine line can be performed.

Some characteristics of the patient can be understood as predictors of a worst prognosis, like being man. Syringomyelia is more frequent to happen in man. And in patients with syringomyelia, specially in those patients who undergone a small increase of occipitopalatine line, is more frequent to develop pseudomeningocele after surgery, which is related with a lower score.

Besides, calculating the CCOS in the appointments after the surgery allows an easier evaluation over time. With the CCOS, the principal information will be assured, and it will be noticed if the patient is getting better or worse.

This study shows that conjugating a good preoperative routine, by calculating occipitopalatine line increase rate, and doing a proper evaluation of the patient after decompressive surgery with the CCOS is beneficial for a correct follow-up of the patients diagnosed with CM1 that need surgery.

Acknowledgments

I thank to Dr. Hermínio Tão for all the support over the last 4 years since we meet each other. My passion for neurosurgery grows every day because of him. I also want to thank him for accepting to coordinate this project, for his continuous assistance, and for all the hours spent at the hospital, not just discussing this work, but also for all the teachings.

I thank Professor Marcos Barbosa for accepting me into his department. I also want to thank him for helping me to improve my knowledge and for the scientific review of the work.

I thank Dr. Ricardo Henriques Pereira for technical support.

Finally, I thank my parents for their unconditional support and encouragement. They have always been by my side during all this process. I thank all my friends for accompanying me on this journey.

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Appendices

Appendix I - Approval of the CHUC's ethics committee

REPÚBLICA PORTUGUESA
SAÚDE

SNS SERVIÇO NACIONAL DE SAÚDE

CHUC CENTRO HOSPITALAR E UNIVERSITÁRIO DE COIMBRA

Comissão de Ética para a Saúde

Visto/ À U.I.D.
para difusão
Dr. Nuno Deveza
Diretor Clínico
C.H.U.C. - EPE

Exmo. Senhor
Dr. Nuno Deveza
Digmº Diretor Clínico do CHUC

SUA REFERÊNCIA	SUA COMUNICAÇÃO DE	NOSSA REFERÊNCIA	DATA
		Nº 368/CES	25-10-2021
		Proc.Nº **OBS.SF.126-2021**	

Estudo Observacional: **OBS.SF.126-2021** *"Cirurgia descompressiva no contexto de Malformação de Chiari tipo 1: uma análise retrospetiva."*
Entrada na UID: 02-07-2021
Entrada na CES: 17-09-2021
Investigador/a/es: António Pinheiro Pinto, Aluno do Mestrado Integrado em Medicina
Coordenador/a/es:
Co-Investigador/a/es: Hermínio José Tão Espírito Santo, Marcos Daniel de Brito da Silva Barbosa
Promotor: Não se aplica
Serviço de Realização: Serviço de Neurocirurgia do Centro Hospitalar e Universitário de Coimbra

Cumpre informar Vossa Ex.ª que a CES - Comissão de Ética para a Saúde do Centro Hospitalar e Universitário de Coimbra, reunida em 20 de Outubro de 2021, após reapreciação do projeto de investigação supra identificado, emitiu o seguinte parecer:

"A Comissão considera que se encontram respeitados os requisitos éticos adequados à realização do estudo, pelo que emite parecer favorável ao seu desenvolvimento no CHUC, com dispensa de consentimento informado".

Mais informa que a CES do CHUC deverá ser semestralmente atualizada em relação ao desenvolvimento dos estudos favoravelmente analisados e informada da data da conclusão dos mesmos, que deverá ser acompanhada de relatório final.

Com os melhores cumprimentos,

A Comissão de Ética para a Saúde do CHUC, E.P.E.

Prof. Doutora Margarida Silvestre
Presidente

CES do CHUC: Prof. Doutora Margarida Silvestre, Enfª Adélio Tinoco Mendes, Dra. Cláudia Santos, Dra. Isabel Gomes, Dra. Isabel Ventura, Rev. Pe. Doutor Nuno dos Santos, Dr. Pedro Lopes, Doutora Teresa Lapa, Dra. Teresa Monteiro

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